Steroid Responsive Acute Recurrent Visual Loss: An Unusual Presentation Of Fibrous Dysplasia

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Citation

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Abstract

We report a case of orbitofrontal fibrous dysplasia that presented with three episodes of acute recurrent visual loss. All the episodes of vision loss were reversed by corticosteroid treatment. Though surgical intervention is generally advocated when fibrous dysplasia is associated with visual loss, corticosteroid therapy may be tried when waiting for surgery or in patients contraindicated for surgery. This is second case reported in literature for successful treatment of fibrous dysplasia associated visual loss with steroid.

INTRODUCTION

Fibrous dysplasia (FD) is a progressive bone disease in which abnormal fibroblast proliferation results in the replacement of normal cancellous bone with an immature fibrous tissue that is poorly mineralized. Orbitofrontal fibrous dysplasia may develop within bone adjacent to the optic canal, grow gradually, and compress the optic nerve leading to visual disturbances ([1,2,3]). We report a case of a patient with unilateral optic neuropathy from fibrous dysplasia that suffered three acute recurrent visual losses which was responsive to steroid therapy. In our knowledge this is second case reported in the literature for successful treatment of fibrous dysplasia with steroid.

CASE REPORT

This 21 years old lady presented with history of painful visual loss of left eye to fingers counting from 1 meter one year back. She was seen by an Ophthalmologist and treated with intravenous (IV) Methylprednisolone for 5 days and regain her vision to 100% (6/6) on fourth day of treatment. Again after 3 months she developed visual blurring with visual acuity of 6/60 in left eye that was improved to 6/9 with oral steroid for 2 months. She presented to us with complain of progressive blurring of vision of left eye since 7 days. But she denied any symptoms such as proptosis, pain in the eyes and diplopia. In addition, she had no history of skin lesions, precocious puberty or other endocrine abnormalities. The patient had no other significant past medical history. General physical and systemic examinations were all normal. She can count only fingers from I meter distance. There was an afferent pupillary defect in the left eye. Slit-lamp examination was unremarkable. Funduscopy showed a normal optic disc. Goldmann visual field testing showed a cecocentral scotoma in the left eye and retrobulbar neuritis was diagnosed. Her blood routine and biochemical examination was normal. Her x-ray chest, routine urine examination, antinuclear antibody titre, anti-double stranded deoxyribonucleic acid titre and thyroid function test were normal. Visual evoked potentials (VEP) using pattern reversal showed significant delay in the P 100-wave latency on the left side. The latency of the right side was normal. Her cerebrospinal fluid routine examination was normal. In view of clinical history and examination supported by VEP and field examination, the patient was diagnosed as case of optic neuritis and started IV Methylprednisolone 1 gm / day for 5 days. After 4 days of treatment, her visual acuity had improved to 12/6 with resolution of the afferent pupillary defect and visual field improvement. Magnetic resonance imaging done at day five of IV Methylprednisolone revealed a diffuse thickening with mixed signal changes of the fronto-parieto-temporal bone and the greater wing of the sphenoid bone on the left side with compression of the left optic nerve (fig 1). The patient underwent orbito-cranial reconstruction and unilateral optic canal release using an extradural approach through a left fronto-temporal craniectomy. Histological findings confirmed the lesion to be typical fibrous dysplasia by presence of multiple small and irregular spicules of abundant immature bone without any osteoblastic lining separated by moderately cellular collagen tissue without any cellular atypia (fig 2). She recovered completely

one month after the operation. Patient was stable over 1 year of follow-up. Postoperative follow-up did not reveal disturbances in visual function, extraocular motility, or evidence of cerebrospinal fluid fistulas.

Figure 1

Figure 1: Magnetic resonance imaging (T1 sequence) showing a diffuse thickening with mixed signal changes of the fronto-parieto-temporal bone and the greater wing of the sphenoid bone on the left side with compression of the left optic nerve.

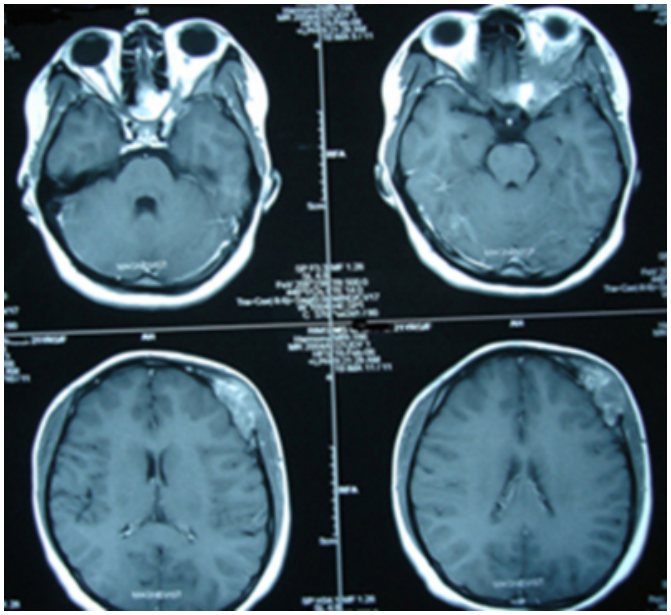
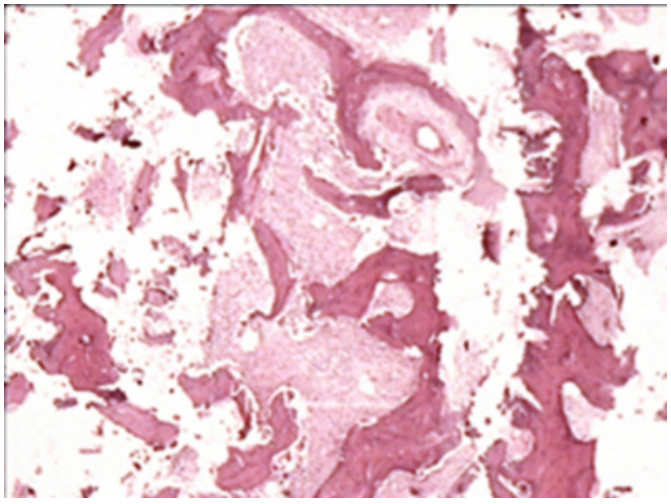


Figure 2

Figure 2: Histological examination showing the presence of multiple small and irregular spicules of abundant immature bone without any osteoblastic lining separated by moderately cellular collagen tissue.



DISCUSSION

Fibrous dysplasia is a rare condition and is reported in 1:4000 - 10,000 people ([4]).

In fibrous dysplasia symptoms arise due to the expansion and impingement of bone on adjacent structures. The most common symptom is swelling or deformity of the affected site ([5]). In our patient, the frontal bone and sphenoid wings were involved with compression of the left optic nerve. Visual impairment is the most feared and potentially debilitating among all the sequele of fibrous dysplasia. Visual loss also represents the most common neurological complication of fibrous dysplasia affecting the skull ([6]). Patients afflicted with this disorder typically present with exophthalmos, displacement of the globe, abnormalities of extraocular motility, and visual impairment. Visual impairment in patients with fibrous dysplasia may include, alone or in combination, perturbances in color vision, central and peripheral field defects, and afferent pupillary defects. It may occur in a chronic and progressive manner or may manifest in an acute condition ([3,7,8]). Acute visual loss is probably the result of the vascular event, affecting the ophthalmic artery and can also result from associated pathologies like mucocele or aneurysmal bone cyst around the vicinity of the optic canal. Our patient presented with acute recurrent visual loss that may be related to vascular event, as there was no visible associated pathology and hemorrhage in the tumor. Various other cause of visual impairment includes optic nerve compression related to optic canal stenosis resulting from fibrous dysplasia, exophthalmos-induced optic nerve traction and spontaneous hemorrhage. Michael and colleagues reviewed 20 cases of fibrous dysplasia with visual loss and attributed 4 cases of visual loss to optic canal stenosis, six patients to cystic fibrous dysplasia, four to hemorrhagic lesions, four to mucoceles and two to aneurismal bone cysts.

Although fibrous dysplasia–induced visual impairment is certainly well documented, its indications for treatment are controversial. The establishment of appropriate therapy for this condition has been hindered by its relative rarity and by the paucity of quality reports in which adequate documentation, including careful imaging studies, is presented. Optic nerve decompression is indicated for patients with visual deterioration. However, in patients with normal vision appropriate management of fibrous dysplasia around the optic nerve remains controversial. Conservative management may be indicated in patients with preserved vision provided visual and radiological assessment done meticulously at each visit. Radiological data, of course, should be interpreted in light of clinical correlations.

Corticosteroid agents have also been used in the care of patients with fibrous dysplasia successfully as a temporizing measure in patients with acute visual loss ($_{10}$). Long term or repeated use of corticosteroid in recurrent visual loss had not been reported so far. Our patient presented with previous history of twice visual blurring which was treated successfully with steroid in both occasion. She presented to us with third episode of visual loss after almost one year after the first episode. Clinical data and preliminary investigation before imaging study was performed, diagnosis of optic neuritis was done and treated with intravenous Methylprednisolone and shown good improvement. Later on only after imaging study was performed the diagnosis of fibrous dysplasia was made. The patient was then subjected to surgical decompression. This is first case to use corticosteroid repeatedly for recurrent visual loss in a case of fibrous dysplasia. Thus a trial of corticosteroid therapy may be beneficial in cases of acute vision loss due to fibrous dysplasia. Further studies needed to confirm usefulness of steroid in fibrous dysplasia. However long-term nonoperative management of visual loss due to fibrous dysplasia with corticosteroid alone may be dangerous as sudden visual function deterioration leading to complete and permanent dysfunction may occur. Surgical intervention may be always advisable in cases of significant, acute, and/or progressive visual impairment is documented by serial clinical and radiological examinations.

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